Abstract  A case of Adams-Oliver syndrome in a Kuwaiti boy with a minor distal limb anomaly is described. This represents a sporadic occurrence of the syndrome, which was treated conservatively in view of the extensive scalp defect. The wound healed completely over a period of 11 months with non-adherent moist dressings. The possible etiologic factors, various clinical presentations, genetic transmission, and the modes of treatment available have been reviewed. We believe this is the first case to be reported in the Middle East.

Key words  Congenital scalp defect

Introduction

Congenital absence of skin and its appendages is a rare congenital anomaly. It can occur in any region of the body. It may be present in varying forms and may be associated with other congenital anomalies. Adams and Oliver in 1945 [1] described the association of cutis aplasia with distal limb anomalies. Since then, 300 cases have been described in the literature [23,33]. The importance of this syndrome lies in its variable expression, genetic transmission, associated anomalies, and the mode of treatment [3,4,12]. The majority of cases are autosomal dominant with several reports of familial transmission, a few being sporadic or autosomal recessive [5,6,17]. A case of Adams-Oliver syndrome of sporadic occurrence, associated with congenital distal limb anomaly, and treated successfully with conservative treatment over a period of 10 months is reported. This may be the first report of Adams-Oliver syndrome from the Middle East.

Case report

A baby boy was born as the second child to a 30-year-old Kuwaiti woman at full term by normal vaginal delivery on 9 September 1997. The father was a 32-year-old Kuwaiti and not related to the mother of the baby. There was no familial history of any disease or drug intake during the course of pregnancy. There was no history of scalp or limb defect or any other anomaly in the parents, siblings, or near relatives. The karyotype was 46XY with normal details. The birth weight of the baby was 3276 g, the length and head circumference at the 50th percentile, and Apgar scores were 9 and 10 at 1 and 5 min. The baby had an extensive scalp defect involving the whole vertex measuring 14×14 cm; it was devoid of skin, hair and underlying bone and was covered by a parchment-like membrane (dura) and some blood collected over it (Fig. 1).

There was fusion of the toes of both feet with hypoplasia and absence of phalanges of the left 3rd and 4th toes (Fig. 2). The upper limbs were normal. Detailed physical examination did not reveal any other defect. A skigram of the skull revealed a defect of the same size (Fig. 3) as the skin lesion. Cranial CT scan, echocardiography, and abdominal ultrasound did not reveal any abnormality of the brain, heart or abdomen, respectively. Neurosurgical consultation was also obtained. The scalp lesion was treated conservatively with local non-adherent dressings and the application of Bactroban ointment, which was changed every other day.

The baby was discharged from the hospital after 3 weeks with an arrangement to follow-up at the plastic surgery clinic on a regular basis. The scalp defect started to gradually heal from the periphery to form a thin epithelium; the skull defect also reduced in size. Over a period of 11 months, the wound healed almost completely except for a small portion of scalp posteriorly (Fig. 4). At the end of 1 year, examination of the baby revealed normal growth according to age in relation to weight, head circumference and length.

Discussion

Congenital cutis aplasia in the scalp was first described by Campbell [5] in 1826; the same condition in the extremities was first described by Cordon [6] in 1967. Since then several reports have appeared in the literature either as isolated anomalies or in association with other congenital defects of the body [16,18]. The maximum number of cases (86%) are found to involve the scalp. These defects of the scalp may vary from small to large skin defects associated with absence of the underlying
Fig. 1 Extensive scalp defect covered by a parchment-like membrane
Fig. 2 Incomplete syndactyly of toes of both feet with absence of phalanges of the left third and fourth digit
Fig. 3 Central bony defect of the scalp
Fig. 4 Almost complete healing of the wound